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Case Report

Merkel cell carcinoma of the breast:
Report of a case and review of the literature

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CASE REPORT A 52-year-old lady presented to A&E in November 2001 with a history of an injury to the upper outer quadrant of the left breast several weeks previously. On examination she had a markedly abnormal left breast. A fungating tumour occupied the upper outer quadrant of the breast with the rest of the breast swollen, oedematous and very firm. Ultrasound scan showed no focal mass but the entire breast was abnormal with areas of low density and interstitial oedema. An Ultrasound guided fine needle aspirate revealed malignant cells. The main differential diagnosis lay between a poorly differentiated carcinoma and a high grade lymphoma therefore a core biopsy was obtained. The core biopsy suggested a small cell carcinoma arising primarily within the breast. A staging CT scan of chest and bone scan revealed liver and bony metastases and chemotherapy was commenced.

Following chemotherapy the breast lesion was clinically smaller and the breast softer, but the lesion was ulcerated and causing a lot of distress. A repeat CT scan confirmed the breast mass had reduced significantly from 10cm to 5cm however liver metastatic deposits had increased in size from a maximum diameter of 3cm to a maximum of 4.5cm. MRI scan of spine revealed extensive bone metastases throughout cervical, dorsal, lumbar spine and sacrum. To improve her quality of life she elected to undergo a left mastectomy and axillary node clearance. At operation the tumour was very vascular and she required a blood transfusion post-operatively. Pathology of the tumour was very unusual for a primary breast carcinoma and further immunohistochemical stains revealed a *Merkel* cell carcinoma. This lady's clinical pattern of a 10cm breast lump with a small skin lesion was extremely unusual.

During her post-operative period she developed low back pain which appeared to settle with analgesia and she was re-admitted soon after discharge with lower limb paralysis. MRI scan revealed an extra-spinal cord deposit. She had radiotherapy and chemotherapy with no return of power in her legs. She subsequently deteriorated very rapidly and sadly passed away.

DISCUSSION

Merkel cell carcinoma (MCC) is a rare cutaneous neoplasm first described by Toker in 1972 as 'trabecular carcinoma'. Merkel cells are believed to be primary neuroendocrine cells found within the basal layer of the dermis. It has been shown to be a highly aggressive and lethal tumour, comparable with small-cell lung cancer and melanoma in its behaviour with regards to recurrence, metastatic spread, and mortality. It is a disease of the elderly, with an average age at the time of diagnosis of 69 years. It affects primarily the sun exposed areas of the skin especially head and neck. Little is known about specific aetiologic factors in the pathogenesis of MCC, however, like melanoma, ultraviolet radiation appears to be a significant factor with respect to both its anatomic and geographical distribution.

Patients typically present with a reddish blue, firm, non tender, nodular mass that has grown

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rapidly over a few weeks to months and which may ulcerate. Due to its non-specific appearance diagnosis is generally not made before biopsy and is then based on immunohistochemical staining for cytokeratin 20. Diagnostic evaluation includes CT imaging, octreotide and PET scans.

Treatment is by wide surgical excision of the primary tumour and chemotherapy, followed by radiotherapy for patients with advanced local and regional spread. Typical spread is to lymph nodes, liver, lung and bones. MCC has a local recurrence rate of 40-44% after primary treatment, a 55% rate of lymph node metastases, and a 34-49% rate of distant metastases with the majority of recurrences appearing within the first six to 12 months after initial diagnosis. Due to the rare occurrence of MCC, no prospective clinical studies assessing initial surgical therapy, radiotherapy, or chemotherapy have been performed.

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